



ABDOMEN IS STILL A PANDORA'S BOX - PREGANGRENOUS BOWEL IN CONGENITAL MALROTATION!

General Surgery

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ABSTRACT

Introduction: Acute abdomen is a condition where the possibility of different organ systems can be involved. Even if you identify the system the clinician has to identify the possible pathology. Th task is difficult even with a thorough history and clinical examination. Here we report a case which would shed light on clinical decision making. **Case Report:** A 20 years old student presented with lower abdominal pain for the past 5 hours to the emergency department. She had guarding and tenderness below the level of umbilicus. Xray, Ultrasound were normal, CT abdomen was which showed some loops of small bowels with malrotation but nothing else. We did Diagnostic laparoscopy that was converted into a laparotomy which showed a pregangrenous segment of ileum twisted around its mesentery which was untwisted. The rest of the small bowel was in a peritoneal pouch with a narrow entry band which was cut to release the bowel. The whole small bowel was traced and flimsy adhesions between them released. The large intestine was normally placed. The pregangrenous bowel recovered in the meantime and a resection was avoided. **Conclusion:** Imaging investigations form only a portion of the algorithm of managing the acute abdomen. If they don't corroborate with the clinical findings, a surgeon's timely decision to operate can avert dreadful complications.

KEYWORDS

Acute abdomen, Malrotation, Pregangrenous, Laparoscopy, laparotomy

INTRODUCTION:

Abdomen has so many systems and organs ranging mainly from the gastrointestinal system, urinary system and reproductive system. Pathology of them can produce a wide spectrum of symptoms and signs often overlapping with each other causing diagnostic dilemmas. Of them all acute abdomen is a section apart where the clinician has to make a diagnosis at the earliest and act upon it swiftly to avert major complications. A poorly managed acute abdomen patient can cause severe morbidity and sometimes mortality. So treating an acute abdomen should not be taken lightly and if necessary we should order appropriate investigations to corroborate or clear the doubts of a working clinical diagnosis so that precious time is not lost.

Acute abdomen in a woman often complicates the equation because of the reproductive system pathologies coming into play. Even though a lower abdominal pain is most often due to acute appendicitis (if it localises in the Right iliac fossa) in both the genders, ureteric colic to be considered if it radiates from the flank. In women, Cystitis, ectopic pregnancy, twisted ovarian cyst, degeneration of fibroid, endometriosis and pelvic inflammatory disease can compound the differential diagnosis. Bowel pathologies like diverticulitis, volvulus, acute intestinal obstruction due to various causes can also be the cause. In India abdominal Tuberculosis should also be considered strongly as the incidence is fairly high. With this background we will discuss a case of acute abdomen that was unique.

CASE REPORT

Acute abdomen still causes diagnostic challenges and sometimes nightmares due to incorrect decisions. A 20 years old college going, unmarried girl presented to the emergency department in the wee hours with acute lower abdominal pain that started 5 hours before. She said that the pain was severe, excruciating located in the lower abdomen below the level of umbilicus but was unable to pinpoint the type or location correctly. She had nausea but did not vomit. She never had a similar pain in the past. Her menstrual was elaborated to rule out ovarian causes which were 4/30 cycles and was regular. Her last menstrual period was 10 days ago and was uneventful. On examination the patient was afebrile, no pallor but was dehydrated which we started to correct by establishing lines. The patient was still holding her abdomen without tossing up in the bed almost ruling out

urolithiasis pain. On examination of the abdomen, it was flat but the lower quadrants did not move with respiration. The lower quadrants below the umbilicus had guarding, severe tenderness and rebound tenderness. The bowel sounds were moderately increased. The digital rectal examination was normal.

Clinically there was peritonism features and we suspected acute appendicitis or an ovarian pathology like twisted ovarian cyst or an ectopic pregnancy as the differential diagnosis. We did Urine pregnancy test to rule out an unlikely pregnancy but it was negative. So, we proceeded to do X-ray Abdomen which was normal, Emergency ultrasound abdomen was taken which too turned out to be normal. Since the clinical suspicion of a pathology was high, we went ahead with Contrast Enhanced Computed Tomography Scan of the abdomen (CECT) which was suspicious of a malrotated small bowel but that does not explain the peritonism features. In the meantime, the abdominal pain partially settled but the signs did not. So, we planned for surgery – Diagnostic Laparoscopy and proceed with proper consent.

Diagnostic laparoscopy: Appendix was visualised and it was absolutely normal. A foot of small bowel lying in Left iliac fossa was severely congested and in a 'Pregangrenous' state [Figure 1,2]. So, a decision was made to convert the surgery into open and we did midline laparotomy. One feet of terminal ileum was congested, pregangrenous and twisted around its mesentery [Figures 3]. We untwisted it and gave 100% oxygen. Warm saline towels were placed over it to increase the blood flow and salvage that segment of ileum. Meanwhile we were trying to figure out the malformed anatomy.

The rest of the small bowel was lying inside a peritoneal pouch with a small opening as entry which had a thickened neck as a band [Figure 4,5]. We released the band and opened the peritoneal sac to deliver the rest of the small bowel which was collapsed and healthy [Figure 6]. In the meantime, the congested ileal segment showed signs of recovery and dropped the idea of resecting it [Figure 7,8]. We traced the small bowel right from Duodeno-Jejunal flexure till Ileo-Caecal Junction and released few more flimsy adhesions. The large intestine was in place. We performed an appendicectomy to avoid future diagnostic dilemmas. We closed the abdomen in layers. The patient recovered well and was doing fine after 6 months follow up.



Figure 1: In Diagnostic Laparoscopy, a segment of small bowel looks congested and Pregangrenous

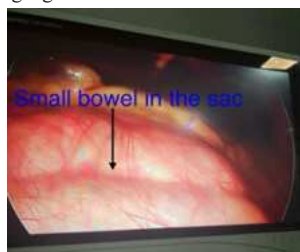


Figure 2: In Diagnostic Laparoscopy, there was a large peritoneal sac seem to contain major share of the bowel

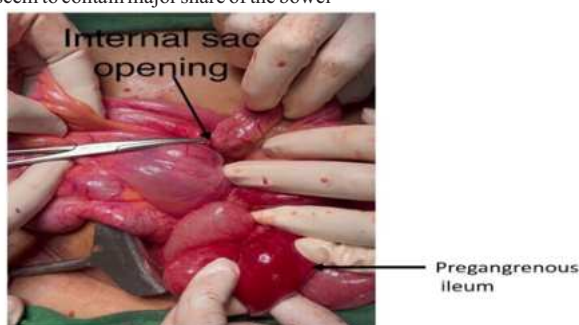


Figure 3: The peritoneal sac had a narrow neck and the pregangrenous bowel in the lower part of the picture

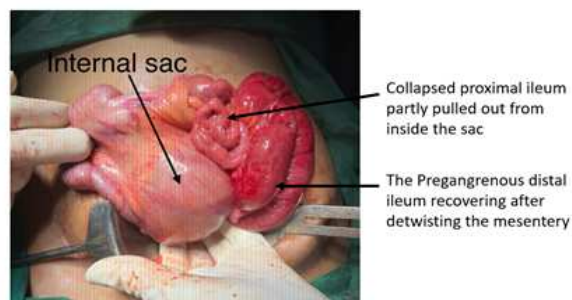


Figure 4: The terminal ileum twisted around the mesentery while the rest of the small bowel was in inside a peritoneal sac



Figure 5: The constriction band being released



Figure 6: The sac was splayed open and the rest of the small bowel delivered out (all lying in the right side of abdomen inside the sac)

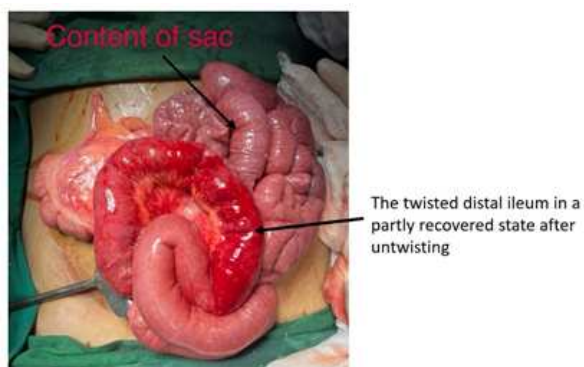


Figure 7: The untwisted portion of ileum given 100% oxygen and seen in the recovery phase



Figure 8: The untwisted ileum has recovered completely.

DISCUSSION:

Malrotation of the gut is generally regarded as a paediatric pathology with the majority of patients presenting in childhood. The diagnosis is uncommon in adults, which often leads to delay in diagnosis and treatment. So therefore, malrotation to be considered as a differential diagnosis when dealing even adults with abdominal symptoms. Midgut malrotation is a congenital anomaly in the embryological development of naturally occurring intestinal rotation. It has been estimated to occur approximately 1 in 500 live births [1]. However, the true incidence is difficult to estimate as a substantial number of cases will go undetected throughout life. The majority of the complications associated with midgut malrotation present in the first month of life amounting to 60-85% of cases [1,2]. Adult midgut malrotation is very rare and its incidence reported by few studies are between 0.0001% and 0.19% [3,4]. Most adult diagnoses of midgut malrotation are made in asymptomatic patients incidentally either on imaging studies for unrelated conditions or at operations for other reasons. However, there are a small proportion of adults especially young adults who may present with acute or chronic symptoms of intermittent and recurrent abdominal pain or intestinal obstruction. The diagnosis in this age group has immense difficulty, especially because the typical presentation is with non-specific abdominal symptoms and the Surgeons might have low index of suspicion of malrotation in the initial evaluation of adult patients with abdominal pain.

Midgut malrotation is considered as any deviation from the normal 270-degree counterclockwise rotation of the gut that occurs in embryonic development. During week 4 of foetal development, the embryonic gut, develops vascular pedicles to be divided into the foregut, midgut and hindgut based on the blood supply. The midgut is supplied by the superior mesenteric Artery (SMA) and by the fifth week it begins to rapidly elongate and a temporary herniation into the umbilical cord with return to the abdominal cavity about 4 to 6 weeks later. During this process, the midgut undergoes a 270-degree counterclockwise rotation around the pedicle of Superior Mesenteric Artery. The progressive reduction of the midgut herniation makes the duodeno-jejunal flexure reduce first and lie to the left. The distal small bowel then follows to lie on the right of the abdominal cavity. The base of the small bowel mesentery then fuses with the posterior peritoneum diagonally, from the ligament of Treitz to the caecum in the Right iliac fossa completing the process by the 11th week of foetal life [4-6]. The pathology of this process, categorises into nonrotation and incomplete rotation [7]. Incomplete rotation refers to a wide range of partial

rotational anomalies possible. Reversed rotation is also possible but quite rare [9]. Nonrotation is noted mostly in the older population [8]. When Malrotation occurs it not only results in the malposition of the bowel but also in the malfixation of the mesentery. The normal broad mesenteric attachment becomes a narrow pedicle that predisposes to the complication of midgut volvulus. Internal hernia associated with the abnormal peritoneal fibrous bands (of Ladd) that attach to the right colon is another complication of malrotation seen in adults.

There are two patterns of presentations of gut malrotations, acute and chronic [5,10,12]. Chronic presentation is more common in adults. This is characterised by intermittent nonspecific abdominal pain, nausea and vomiting over several months or years. This emphasizes the need for a low threshold to suspect midgut malrotation, when investigating intermittent and varying abdominal symptoms in an otherwise healthy young adult [5,10]. A study reported that 6 of 12 patients in their series presented with chronic intermittent abdominal symptoms [10]. Diagnostic delays eventually happen in these patients and these symptoms need to be properly investigated. Dietz et al studied 10 adults with small bowel obstruction due to intestinal malrotation and found that 5 adults presented with chronic features and the duration of symptoms sometimes extended to even 30 years [5]. The other group of adults typically present with acute symptoms like acute abdomen or bowel obstruction. This was the case in our patient. A study reported that 48.5% of the thirty-three patients presented with an acute abdomen [11]. Acute abdomen may be due to volvulus of the midgut or ileocaecum, reported as the most common cause of bowel obstruction in adults with gut malrotation. Other causes may be related to internal herniation by Ladd's bands. The unusual intestinal anatomy results in atypical signs and symptoms in some. Several studies have reported atypical presentations before discovering gut malrotation with abnormal location of the bowel at surgery [5,10].

Conventional plain X-rays are neither sensitive nor specific in the diagnosis of gut malrotation. Ultrasound might help with findings of midgut volvulus include duodenal dilatation with distal tapering and fixed midline bowel and mesentery twisted around the SMA axis. These features classically present as the 'whirlpool' sign [12]. Colour Doppler Ultrasound may reveal malposition of the SMA [10,13,14]. However, the gold standard for diagnosing gut malrotation would be an upper gastrointestinal (UGI) contrast study, especially in the paediatric age group [5,13]. This will elucidate that the duodenum and duodenojejunal flexure located to the right of the spine. However, contrast study reports may be nonspecific and a normal study does not completely exclude the presence of a gut malrotation [10,13]. CT scan with contrast is increasingly preferred as it is convenient and faster and also gives more details with an accuracy of 80% [14-16]. CT scan show the anomalous position of SMA, and also the abnormal location of the bowel.

CT scan findings of gut malrotation and small bowel obstruction without volvulus, may also show internal herniation secondary to Ladd's bands. Symptomatic midgut malrotation requires emergent surgical intervention although the management of asymptomatic patients is debatable. The surgical management of intestinal malrotation was first described by William Ladd in 1936 [6] and this remains the mainstay of treatment. Choi et al [17] reviewed 177 patients over a 35-year period and found that asymptomatic patients had a low risk of intestinal volvulus and therefore advised that elective surgery is not necessary but close follow-up alone can be done. However, the other school of thought is that all patients with intestinal malrotation should undergo surgical correction as it is impossible to predict who will develop life threatening complications [18]. Several studies though with small numbers have recommended that elective Ladd's procedure should be performed in all patients [5,10,11]. Having said that, the operative decision should be based on the presentation and after weighing the potential risk-benefit ratio.

Midgut malrotation may be quiescent in adults with subtle symptoms but if there is volvulus or twisting of the part of the gut as in our case it may become life-threatening because of the risk of bowel obstruction and strangulation. Strangulation may cause sepsis and may result in mortality also. Some patients may present with a combination of midgut volvulus and internal hernia. Our case was unique that it had a peculiar anatomy with internal herniation into a peritoneal sac and the small part of terminal ileum lying outside had twisted around its axis. This learning point leads us to opine that surgical treatment of asymptomatic malrotation should be considered because that's the only solution to prevent complications.

CONCLUSION:

This case report emphasises the fact that clinical skills and acumen of a Surgeon is critical if the imaging and other adjunct investigations doesn't support the diagnosis. By the decision to operate in time we have prevented a late diagnosis which could have caused gangrene of small bowel, sepsis and a vicious cycle. Malrotation should also be considered as a differential diagnosis in acute abdomen especially in young adults. Abdomen is still a Pandora's box but Surgeons have a better understanding of the box than before.

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